

Case Report

Pudendal Neuralgia Due to Pudendal Nerve Entrapment: Warning Signs Observed in Two Cases and Review of the Literature

Stéphane Ploteau, MD^{1,2}, Claire Cardillac, MSc², Marie-Aimée Perrouin-Verbe, MD¹, Thibault Riant, MD^{1,3}, and Jean-Jacques Labat, MD¹

From: ¹Federative Pelvic Pain Center, Nantes, France; ²Department of Gynecology-Obstetrics and Reproductive Medicine, Centre Hospitalier Universitaire, Nantes, France; ³Maurice Bensaïgnor Multidisciplinary Pain Center, Centre Catherine de Sienne, Nantes, France

Address Correspondence:
Stéphane Ploteau, MD
Department of Gynecology-Obstetrics and Reproductive Medicine
38 Boulevard Jean Monnet
Women's and Children's Hospital,
University
Nantes, France
E-mail:
stephane.ploteau@chu-nantes.fr

Manuscript received: 07-21-2015
Revised manuscript received:
09-05-2015
Accepted for publication:
10-12-2015

Free full manuscript:
www.painphysicianjournal.com

Pudendal neuralgia is a chronic neuropathic pelvic pain that is often misdiagnosed and inappropriately treated. The Nantes criteria provide a basis for the diagnosis of pudendal neuralgia due to pudendal nerve entrapment. The 5 essential diagnostic criteria are pain situated in the anatomical territory of the pudendal nerve, worsened by sitting, the patient is not woken at night by the pain, and no objective sensory loss is detected on clinical examination. The fifth criterion is a positive pudendal nerve block. We have also clarified a number of complementary diagnostic criteria and several exclusion criteria that make the diagnosis unlikely. When pudendal neuralgia due to pudendal nerve entrapment is diagnosed according to the Nantes criteria, no further investigation is required and medical or surgical treatment can be proposed. Nevertheless, a number of warning signs suggesting other possible causes of pudendal neuralgia must not be overlooked. These warning signs (red flags) are waking up at night, excessively neuropathic nature of the pain (for example, associated with hypoesthesia), specifically pinpointed pain, which can suggest neuroma and pain associated with neurological deficit. In these atypical presentations, the diagnosis of pain due to pudendal nerve entrapment should be reconsidered and a radiological examination should be performed. The 2 cases described in this report (tumor compression of the pudendal nerve) illustrate the need to recognize atypical pudendal neuralgia and clarify the role of pelvic magnetic resonance imaging (MRI), as MRI provides very valuable information for the evaluation of diseases involving the ischioanal fossa. The presence of red flags must be investigated in all cases of pudendal neuralgia to avoid missing pudendal neuralgia secondary to a mechanism other than nerve entrapment.

Key words: Pudendal nerve, pudendal neuralgia, Nantes criteria, pelvic pain, pudendal canal, perineal pain

Pain Physician 2016; 19:E449-E454

The diagnosis of pudendal neuralgia due to pudendal nerve entrapment is essentially clinical. After having been neglected and poorly defined for a long time, pudendal neuralgia can now be easily diagnosed in the presence of typical perineal pain, previously considered to be psychogenic due to the absence of organic lesions demonstrated on imaging. In 2008, we described a number of converging arguments in favor of the diagnosis of pudendal nerve entrapment, the most common cause of pudendal neuralgia (1). All physicians concerned

with the perineum should be familiar with this limited number of criteria to avoid misdiagnosis in patients reporting perineal pain. Five criteria are considered to be essential and must be present in order to conclude pudendal nerve entrapment or compressive pudendal neuralgia:

1. Pain in the territory of the pudendal nerve from the anus to the penis or clitoris. This nerve trunk pain is superficial or slightly deeper in the ano-rectal and vulvovaginal areas and distal urethra. This criterion excludes pain in other nerve trunk

territories such as the cluneal, ilioinguinal, or iliohypogastric nerves. It must be remembered that, in men, sacral nerve roots and pudendal nerves innervate the scrotal skin, while the testis (like the ovary), epididymis, and vas deferens are innervated by thoracolumbar nerve roots (autonomic innervation).

2. Pain predominantly experienced while sitting. This criterion is essential, as the nerve is no longer mobile when sitting and is compressed between the sacrospinous and sacrotuberous ligaments. Excessive pressure on the nerve is then responsible for pain. In contrast, sitting on the toilet seat is not painful.
3. The pain does not wake the patient at night. This absence of pain at night is related to the previous criterion. When the patient reports waking up at night, imaging is recommended to exclude nerve compression by a tumor.
4. No objective sensory impairment. The absence of sensory impairment can be explained by anatomical data, as several anatomical territories innervated by several different nerves overlap in this region (pudendal nerve, posterior femoral cutaneous nerve and its cluneal branches, iliofemoral and ilioinguinal nerves). A sensory deficit in the territory of the pudendal nerve is suggestive of a sacral nerve root or sacral plexus lesion.
5. Pain relieved by diagnostic pudendal nerve block. Subject to an irreproachable technique, a positive pudendal nerve block indicates the presence of a pudendal nerve lesion, but does not indicate whether it is specifically due to nerve entrapment. A positive nerve block significantly relieves the pain for the duration of the local anesthesia.

Eight complementary diagnostic criteria suggestive of or compatible with the diagnosis have also been defined:

1. Burning, shooting, stabbing pain, and numbness are sometimes present and are characteristic of neuropathic pain.
2. Allodynia or hyperpathia are also suggestive of neuropathic pain (intolerance of tight clothes and underwear).
3. Rectal or vaginal foreign body sensation. This type of pain has an autonomic connotation and temporary relief of these sensations by anesthetic block of the ganglion impar suggests that they are mediated by sympathetic fibers.
4. Worsening of pain during the day, which is a very

characteristic time-course of pudendal neuralgia due to nerve entrapment.

5. Predominantly unilateral pain, which is characteristic of a nerve trunk lesion. Often experienced in all of the hemiperineum, midline or central pain does not exclude the diagnosis.
6. Pain triggered by defecation, not immediately after defecation but generally after several minutes.
7. Exquisite tenderness on palpation of the ischial spine during digital rectal or vaginal examination. This sign is not specific, but suggestive when unilateral, as many anatomical structures are situated in this area (pudendal nerve, insertions of sacrospinous ligament, ischiococcygeal fibers of levator ani muscles). This tenderness may also correspond to diffuse hypersensitivity.
8. Clinical neurophysiology findings in men or nulliparous women, especially when unilateral. Neurophysiology examination is not indicated in multiparous women because of the high rate of perineal lesions after delivery.

Four exclusion criteria that make the diagnosis unlikely have also been defined:

1. Exclusively coccygeal, gluteal, pubic, or hypogastric pain that does not correspond to the anatomical territory of the pudendal nerve.
2. Pruritus is more suggestive of a dermatological lesion.
3. Exclusively paroxysmal pain is suggestive of a compressive lesion and justifies imaging studies.
4. Imaging abnormalities able to account for the pain. Nerve entrapment cannot be visualized by imaging, but imaging can be useful to exclude other diagnoses. Caution must be observed in the presence of anomalies for which treatment will not modify the pain (arachnoid cysts, etc.).

Several associated signs that do not exclude the diagnosis have also been described, as many patients describe additional symptoms that can sometimes be confusing because they are obviously unrelated to the pudendal nerve:

1. Buttock pain on sitting. This pain is due to myofascial syndromes, as several structures can enter into conflict with the piriformis muscle (pudendal nerve, posterior femoral cutaneous nerve, inferior gluteal nerve, and sciatic nerve). Piriformis muscle spasm can induce pain in the territories of these nerves. Buttock pain can also be related to trigger

- points in the obturator internus muscle.
2. Referred sciatic pain. More or less truncated sciatica is frequently associated with pudendal neuralgia due to a lesion of the posterior femoral cutaneous nerve or sciatic trunk with or without a piriformis or obturator internus syndrome.
 3. Pain referred to the medial aspect of the thigh, territory of the obturator internus nerve. This symptom is explained by the contact of the nerve with this muscle.
 4. Suprapubic pain related to hypertonia of the puborectalis component of the levator ani muscles or to bone hypersensitivity suggestive to a complex secondary pelvic pain syndrome.
 5. Urinary frequency and/or pain on a full bladder. Urinary frequency is often associated with pudendal neuralgia in the absence of bladder dysfunction. These symptoms are due to secondary regional sensitization.
 6. Pain occurring after ejaculation. This symptom, which is perplexing in the absence of infection, is fairly frequently associated with pudendal neuralgia and can be considered to reflect regional sensitization.
 7. Dyspareunia and/or pain after sexual intercourse. The frequency of sexual intercourse is often reduced in patients with pudendal neuralgia, as pain decreases libido. Sexual intercourse is rarely painful, except in the case of vulvodynia. As a result of regional sensitization, these women often experience pain over the hours following intercourse.
 8. Erectile dysfunction. Pudendal neuralgia, affecting a somatic nerve, only has a limited effect on erection, as the main sexual function of the pudendal nerve is sensory (dorsal nerve of the penis or clitoris), but it is also involved in the rigid-erection phase before ejaculation and in the clonic nature of ejaculation. Patients with pudendal neuralgia frequently complain of a feeling of penile numbness, decreased sexual sensations, or even decreased penile rigidity.
 9. Normal clinical neurophysiology, particularly because these examinations only investigate large motor fibers and may not detect selective lesions of small sensory fibers. Pain is also intermittent and positional (2).

The diagnosis of pudendal neuralgia is therefore essentially clinical, in which the patient's clinical history plays a major role. Nevertheless, various warning

signs may suggest other causes of pudendal neuralgia that must not be overlooked. These warning signs are waking up at night, excessively neuropathic nature of the pain (for example, associated with hypoesthesia), specifically pinpointed pain, which can suggest neuroma, and pain with neurological deficit (sensory deficit or motor deficit, for example, associated with incontinence).

In the presence of atypical pain, the diagnosis of pain due to pudendal nerve entrapment should be reconsidered, and a radiological examination should be performed.

METHODS

We report 2 extremely rare cases of endometrial stromal sarcoma and adenoid cystic carcinoma arising in the ischiorectal fossa that were discovered after consultation for pudendal neuralgia. Both tumors compressed the pudendal nerve and presented in the form of atypical pudendal neuralgia.

These patients provided their written informed consent for the procedures, digitization of their medical records, and for use of data related to their medical history

Case 1:

A 31-year-old woman was referred to the multidisciplinary perineal pain clinic in 2014 with chronic perineal pain syndrome. She had initially consulted a gynecologist in 2011 at the time of onset of the pain. Pelvic magnetic resonance imaging (MRI) showed a solid lesion with small cystic components in the anterior part of the ischiorectal fossa. The tumor measured 30x30x20 millimeters. Diagnostic laparoscopic surgery was normal. Exploration of the ischiorectal fossa via a vaginal incision was performed during surgery. A dystrophic area in contact with the levator ani muscle was demonstrated. Open biopsy was performed and histological examination demonstrated endometriosis. The patient's perineal pain remained unchanged after surgery.

The patient was then referred to our department. She reported an unremarkable medical and surgical history. At the initial assessment, she described suggestive clinical symptoms, such as intense neuropathic pain throughout the anatomical territory of the left pudendal nerve. Pain was located in the left labium majus and vagina. She also described an atypical clinical presentation according to the Nantes criteria: pain was not exacerbated in the sitting position and woke

her up at night. Palpation revealed an area of allodynia on the left vaginal wall and a palpable mass in the ischioanal fossa. Pelvic MRI was performed to assess local progression of the mass over 3 years and revealed a persistent and stable tumor (Fig. 1) in the left ischioanal fossa with heterogeneous contrast. The mass presented solid and cystic components. The tumor was adjacent to the external urethral sphincter and pudendal canal, with no



Fig. 1. Pelvic MRI (case 1): axial and sagittal T2-weighted images of the left ischioanal fossa tumor (endometrioid stromal sarcoma).

obvious invasion of adjacent tissues. This tumor may have had a direct compressive effect on the pudendal nerve in the pudendal canal. Computed tomography (CT)-guided biopsy was performed. Histological examination demonstrated a lesion with smooth muscle differentiation, leading to a diagnosis of endometrioid stromal sarcoma or cellular leiomyoma. The mass was removed via a transischial approach. Macroscopic examination revealed a nonencapsulated, mixed lesion with a gelatinous component and a solid component. Histopathological examination confirmed the diagnosis of endometrioid stromal sarcoma with smooth muscle differentiation and expert confirmation by an international center was obtained. The patient reported no persistent pain or dyspareunia at the postoperative visit.

Case 2:

A 68-year-old woman was referred to our unit with left perineal pain presenting neuropathic characteristics in the pudendal nerve territory. The patient reported onset of a mass on the left posterior labium majus after a fall, 3 years prior to referral, which had subsequently remained stable. The unilateral burning, electric-shock-like pain was located in front of the mass, on the left labium majus, and was exacerbated in the sitting position. The patient was woken at night by the pain. The pain was not exacerbated by micturition or bowel movement. Physical examination revealed a painful mass approximately 3 centimeters large, involving Bartholin's gland. Pelvic MRI demonstrated a mass (70x20x30 millimeters) with extension into the anterior part of the left ischioanal fossa; the mass had infiltrated the pudendal canal. The tumor had invaded the left vaginal wall and left side of the external urethral sphincter. Ultrasound-guided percutaneous biopsy demonstrated adenoid cystic carcinoma.

The patient initially refused surgery because of the mutilating nature of the procedure. On the 6-month follow-up pelvic MRI, the tumor had increased in size (70x30x30 millimeters) and the patient then

agreed to surgery. Laparoscopic surgery revealed a tumor adjacent to the obturator internus muscle with no signs of infiltration. Partial left vulvectomy (excision of Bartholin's gland) without lymph node dissection was performed. Histological examination demonstrated adenoid cystic carcinoma arising from Bartholin's gland. The resection margins were positive with extensive perineural invasion, requiring more extensive surgery.

Discussion

Pudendal neuralgia is a form of chronic neuropathic pain (3) that causes significant impairment of the patient's quality of life. It is observed with a gender ratio of 6 women for 4 men, with no differences in terms of the clinical presentation of pudendal neuralgia between men and women. Pudendal neuralgia is often misdiagnosed and inappropriately treated, as many men with pudendal neuralgia are initially diagnosed with chronic prostatitis/chronic pelvic pain syndrome and many women with a diagnosis of vulvodynia actually suffer from pudendal neuralgia. For these reasons, in 2008, we tried to define diagnostic criteria for pudendal neuralgia due to pudendal nerve entrapment. We established a limited number of simple criteria designed to avoid excessive or incorrect diagnosis of pudendal neuralgia. These criteria were validated by a multidisciplinary working party in Nantes and were then approved by the SIFUD PP (Société Interdisciplinaire Francophone d'Urodynamique et de Pelvi-Périnéologie) and are now included in a consensus-based terminology report for female pelvic floor dysfunction (4). These Nantes criteria provide a basis for the diagnosis of pudendal neuralgia due to pudendal nerve entrapment (1). The 5 essential diagnostic criteria are pain in the anatomical territory of the pudendal nerve, worsened by sitting, the patient is not woken at night by the pain, and there is no objective sensory loss on clinical examination. The fifth essential criterion is a positive anesthetic pudendal nerve block.

The presence of these criteria is mandatory to confirm the diagnosis of pudendal neuralgia. In these cases, no further investigation is required and medical or surgical treatment can be proposed.

However, if any criteria are missing or if the patient has an unusual clinical presentation or presents any exclusion criteria, such as exclusively paroxysmal pain, the diagnosis of pudendal neuralgia due to pudendal nerve entrapment cannot be confirmed and MRI is generally required.

These 2 case reports illustrate the need to recog-

nize atypical pudendal neuralgia, as these patients reported unusual symptoms that should alert the physician. In both cases, pain was intensely neuropathic and woke the patient at night. The 2 tumors described in these patients are rare gynecologic malignancies. Primary carcinoma of Bartholin's gland accounts for 0.001% of all female genital tract malignancies; it is a slow-growing tumor with late clinical presentation associated with a risk of recurrence and distant metastasis. Fewer than 100 cases have been reported in the literature to date (5). These tumors are rarely diagnosed in a context of vulvar pain (6). Extrauterine endometrial stromal sarcoma is also an extremely rare neoplasm, accounting for 0.2% of all uterine malignancies. To the best of our knowledge, only a few case reports and a small case series have been described (7,8). A primary endometrial stromal sarcoma in the ischiorectal fossa is also very unusual. Patients with atypical features of pudendal neuralgia, such as waking at night due to pain or no variation of pain in various positions, require radiological assessment to exclude another cause. MRI provides valuable information for the evaluation of diseases involving the pudendal nerve distribution, such as the ischiorectal fossa (9). When there is a doubt about the diagnosis, these radiological examinations should obviously be performed before nerve blocks to avoid dissemination of tumor cells.

The ischiorectal fossa is a rare tumor site. The symptoms caused by tumors of the ischiorectal region, as illustrated by these 2 cases, are not specific to the lesion but can include atypical unilateral pudendal neuralgia. Patients presenting with pudendal neuralgia should be carefully assessed for warning signs to ensure that pudendal neuralgia is not due to a mechanism other than nerve entrapment. In the 2 cases reported here, pudendal neuralgia was caused by a direct compressive effect on the pudendal nerve in the pudendal canal.

Disclaimer

There was no external funding in the preparation of this manuscript.

Conflict of interest

Each author certifies that he or she, or a member of his or her immediate family, has no commercial association (i.e., consultancies, stock ownership, equity interest, patent/licensing arrangements, etc.) that might pose a conflict of interest in connection with the submitted manuscript.

REFERENCES

1. Labat JJ, Riant T, Robert R, Amarenco G, Lefaucheur JP, Rigaud J. Diagnostic criteria for pudendal neuralgia by pudendal nerve entrapment (Nantes criteria). *NeuroUrol Urodyn* 2008; 27:306-310.
2. Lefaucheur JP, Labat JJ, Amarenco G, Herbaut AG, Prat-Pradal D, Benaim J, Aranda B, Arne-Bes MC, Bonniaud V, Boohs PM, Charvier K, Daemgens F, Dumas P, Galaup JP, Sheikh Ismael S, Kerdraon J, Lacroix P, Lagauche D, Lapeyre E, Lefort M, Leroi AM, Opsomer RJ, Paratte B, Prévinaire JG, Raibaut P, Salle JY, Scheiber-Nogueira MC, Soler JM, Testut MF, Thomas C. What is the place of electroneuromyographic studies in the diagnosis and management of pudendal neuralgia related to entrapment syndrome? *Neurophysiol Clin* 2007; 37:223-228.
3. Ploteau S, Labat JJ, Riant T, Levesque A, Robert R, Nizard J. New concepts on functional chronic pelvic and perineal pain: Pathophysiology and multidisciplinary management. *Discov Med* 2015; 19:185-192.
4. Haylen BT, de Ridder D, Freeman RM, Swift SE, Berghmans B, Lee J, Monga A, Petri E, Rizk DE, Sand PK, Schaer GN. An International Urogynecological Association (IUGA)/International Continence Society (ICS) joint report on the terminology for female pelvic floor dysfunction. *Int Urogynecol J* 2010; 21:5-26.
5. Yang SY, Lee JW, Kim WS, Jung KL, Lee SJ, Lee JH, Bae DS, Kim BG. Adenoid cystic carcinoma of the Bartholin's gland: Report of two cases and review of the literature. *Gynecol Oncol* 2006; 100:422-425.
6. Finan MA, Barre G. Bartholin's gland carcinoma, malignant melanoma and other rare tumours of the vulva. *Best Pract Res Clin Obstet Gynaecol* 2003; 17:609-633.
7. Masand RP, Euscher ED, Deavers MT, Malpica A. Endometrioid stromal sarcoma: A clinicopathologic study of 63 cases. *Am J Surg Pathol* 2013; 37:1635-1647.
8. Kim L, Choi SJ, Park IS, Han JY, Kim JM, Chu YC, Kim KR. Endometrial stromal sarcoma of the small bowel. *Ann Diagn Pathol* 2008; 12:128-133.
9. Hoeffel C, Crema MD, Azizi L, Lewin M, Monnier-Cholley L, Arrivé L, Tubiana JM. Magnetic resonance imaging of the ischiorectal fossa: Spectrum of disease. *J Comput Assist Tomogr* 2007; 31:251-257.